Two-Dimensional Echocardiographic Recognition of Papillary Muscle Fibrosis in Pediatric Patients*

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Two-dimensional echocardiographic examinations were performed in seven infants with anomalous origin of the left coronary artery from the pulmonary artery (ALCA), in one infant with myocardial infarction and left main coronary obstruction, and in eight with critical valvar aortic stenosis (AS). Comparative qualitative echo density assessment demonstrated that each infant had a marked increase in the echo density of one or both left ventricular papillary muscles. This increased echo density was considered to represent fibrosis and scar formation as a result of ischemia and infarction. Pathologic proof of excessive fibrosis of the papillary muscles was obtained in three cases. In an additional case, calcification of the papillary muscle was noted on fluoroscopic examination. The echocardiographic appearance of papillary muscle fibrosis provides a useful indicator of severe subendocardial ischemia in patients with either critical AS or ALCA.

The acoustic properties of the myocardium are altered by the presence of myocardial scarring from infarction or diffuse fibrosis. This has been assessed qualitatively as an increase in the echo density of the affected areas of myocardium. Quantitative assessment of myocardial fibrosis has recently been accomplished by establishment of a color display format which corresponds to the intensity of the echocardiographic image. Two-dimensional echocardiographic studies in adult patients have suggested papillary muscle fibrosis and dysfunction after a myocardial infarction. One recent study of adults described the echocardiographic recognition of fibrosis and calcification of the left ventricular papillary muscles.

Five infants were seen with extremely echodense papillary muscles which were akinetic and associated with mitral insufficiency. Three of these patients had documented anterolateral myocardial infarctions and increased echo density of the anterolateral papillary muscle. Among the latter, two cases were as a result of anomalous origin of the left coronary artery from the pulmonary artery (ALCA) while one was associated with stenosis of the left main coronary artery and diffuse hypoplasia of this structure. The remaining two patients had critical aortic valvar stenosis (AS) with involvement of both the anterolateral and posteromedial papillary muscles. Subsequently, the two-dimensional echocardiograms were examined from ten surviving patients with either critical AS or ALCA and from one infant with ALCA who died. This was done to further evaluate the echocardiographic sign of papillary muscle scarring and dysfunction. Each of these lesions is well known to be associated with pathologic evidence of papillary muscle fibrosis or infarction.

Material and Methods

Review of patient files (1976 to 1981) identified seven infants with ALCA, eight with critical AS, and one with left main coronary artery occlusion. All 16 patients underwent cardiac catheterization and angiography. Two patients were aged 2 to 2.5 years; the remaining 14 were less than 0.5 years.

All patients had two-dimensional echocardiographic examinations utilizing a two-dimensional sector scanner (Advanced Technology Laboratory [ATL]) with either a 3.0 or 5.0 MHz transducer. The images were obtained in standard echocardiographic planes and displayed according to accepted standards.

Great care was taken to view the papillary muscles with an appropriate setting of the gain attenuation. If the attenuation is too high, then differences in the echo density of the abnormal papillary muscles as compared with the adjacent normal myocardium may not be apparent. Similarly, if the gain attenuation is too low, then all of the structures (normal myocardium as well as myocardial scar tissue) may appear echo dense, suggesting a false positive diagnosis of papillary and diffuse muscle fibrosis. Therefore, the gain attenuation was adjusted as high as possible while still allowing resolution of the papillary muscles and adjacent myocardium and endocardial-cavity interface (Fig 1).

All 16 patients had serial ECGs which were examined for evidence of myocardial hypertrophy, ischemia, or infarction.

Results

Fourteen of the 16 patients had cardiac symptoms with severe congestive heart failure and diagnosis in the newborn or early infancy age period. Direct histologic evidence of extensive fibroelastosis of the

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FIGURE 2. Cross-sectional echocardiogram, ft ventricle demonstrates echo-dense papillary muscles (arrows) in patient who underwent surgery for critical aortic stenosis two years previously. Abbreviations as in Figure 1.

ECG and thallium scan and ascribed to obstruction of the left main coronary artery demonstrated by selective coronary arteriography. Fluoroscopic examination of this infant demonstrated calcification in the region of the anterolateral papillary muscle.

The infants with critical AS had an aortic valve pressure gradient ranging from 30 to 100 mm Hg (mean = 65 mm Hg). Angiographic evidence of mitral insufficiency was present in 11 of 16 patients (two severe, three moderate, and six mild) at the initial study.

All patients had abnormal ECGs, demonstrating either an anterolateral myocardial infarction (eight patients) or left ventricular hypertrophy with strain in anterolateral papillary muscles was obtained in two infants with ALCA who died and in one infant with ALCA who required mitral valve replacement for severe mitral insufficiency. A fourth infant had a remote lateral myocardial infarction documented by ECG and thallium scan and ascribed to obstruction of the left main coronary artery demonstrated by selective coronary arteriography. Fluoroscopic examination of this infant demonstrated calcification in the region of the anterolateral papillary muscle.

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dense papillary muscles have not been encountered in over 1,000 other echocardiographic studies during the past year. Until quantitative measurement of regional amplitude of ultrasound reflected from intracardiac structures becomes more widespread, it is suggested that the presumed diagnosis of papillary muscle fibrosis can be made in the proper clinical setting after careful initial attenuation settings when markedly echo-dense papillary muscles are demonstrated in at least two imaging planes (referenced to the surrounding myocardium and pericardium).

There is ample data to support the concept of papillary muscle infarction and fibrosis in pediatric patients with critical AS or ALCA. Noren et al. reported necropsy data from four infants with ALCA and mitral insufficiency. All exhibited fibrosis, scarring, and foci of calcification within their posteromedial papillary muscles. In three of four cases, the anterolateral muscles were also involved, although less extensively. Moller et al. reported autopsy findings from 11 infants with critical AS who died at three days to six months of age. In each case, atrophy and fibrosis were present in one or both papillary muscles. The more severe involvement was usually in the posteromedial papillary muscle. In two instances, calcification of the posteromedial papillary muscle was demonstrated. Pathologic confirmation of papillary muscle fibrosis was possible in two patients, surgical confirmation in one patient who required mitral valve replacement, and fluoroscopic evidence of calcification in the region of the anterolateral papillary muscle was present in one infant.

In the patients with ALCA, myocardial ischemia and often myocardial infarction is secondary to a "steal" effect of reverse coronary flow from the right coronary via collaterals to the left coronary artery and then into the pulmonary artery. The infants with critical AS have subendocardial ischemia and infarction on the basis of coincident increased myocardial oxygen requirements and relative impairment of coronary flow to the left ventricular subendocardial muscle. 11

**Conclusions**

The distinctive echocardiographic appearance of papillary muscle fibrosis would appear to be quite frequent in patients with ALCA or critical AS. Although this localized fibrosis may of itself not cause any major clinical problem, it may be a useful marker of more widespread subendocardial ischemia and reflects the precarious coronary perfusion of the papillary muscles in these two conditions.

**Acknowledgment:** We acknowledge the assistance of Ms. Melanie Gevitz in the preparation of this manuscript and the technical assistance of Sue Pajcic.

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